Multiple sclerosis, vision problems and visual impairment interventions

Information Monitoring Summary

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Summary

Multiple sclerosis (MS) is a degenerative disease leading to progressive demyelination of the central nervous system. The most common form is characterized by a cycle of relapses and remissions, with defined attacks, followed by full or partial remissions [8]. Over time, the body’s capacity to repair myelin may diminish; neurological damage no longer reverses itself and permanent lesions remain [8]. MS presents a multitude of signs and symptoms, among them motor and vision impairments; cognitive and psychological problems may also be present [5, 6, 15, 22, 26].

Optic neuritis is often one of the earliest symptoms of MS. The resulting visual incapacity in the affected eye is typically caused by central scotoma resulting in reduced visual acuity and contrast sensitivity as well as visual field impairments [6, 10, 18]. In most cases, spontaneous recovery of vision occurs with visual acuity of at least 20/40 after the recovery period, which usually lasts 6 to 12 weeks [19]. However, visual impairment may persist. Approximately half of individuals with MS experience a recurrence of optic neuritis during the 10 years following the initial episode [19].

As they grow older, persons with MS present increasingly impaired colour discrimination, which evolves nearly twice as fast as in MS-free individuals [12]. MS may also cause hemianopsia; the extent of recovery varies depending on the extent of the initial visual field impairment [6].

The oculomotor deficit most often associated with MS is internuclear ophthalmoplegia, which leads to diplopia [6]. All types of nystagmus may be encountered in individuals with MS; possible consequences are reading difficulties, oscillopsia (stationary objects appear to be moving), blurred vision or vertigo [6, 24]. Ocular dysmetria and gaze paresis may be present [6, 24].

MS may cause visual impairments that require visual rehabilitation interventions. The person’s needs in terms of magnification, contrast enhancement and enlargement of the visual field should be considered. Also to be taken into account are their physical and cognitive impairments. In conjunction with motor and sensitivity disturbances, vision problems may jeopardize the individual’s safety when they move around, and should be considered accordingly [17, 23, 27].
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1. Multiple Sclerosis

1.1 Etiology and pathogenesis

Multiple sclerosis is regarded as a demyelinating inflammatory disorder of the central nervous system (brain, spinal cord and optic nerve) [11]. Myelin forms a sheath around certain nerve fibres. In the initial stages of the disease, an attack (also called a flare-up or exacerbation) is characterized by inflammation of the myelin along the nerve fibres (axons), which damages or halts nerve impulses [6, 26]. The healing process results in remission (remyelination), which may be full or partial [11, 26]. Over time, the organism’s capacity to repair myelin may diminish; neurological damage is no longer reversed and permanent axonal lesions caused by inflammation then occur [11, 26].

Between 80% and 85% of persons with MS start by presenting a cyclical form of the illness (relapsing-remitting) [26]. Clearly defined acute attacks are followed by full recovery or with sequelae and residual deficit upon recovery [8]. Each attack is followed by full or partial remission, which may last for months or even years [8, 26]. In people with the cyclical form of MS, during the ten years following diagnosis, some 50% will develop the secondary progressive form [15, 26], with progression of the disease with or without occasional relapses, minor remissions and plateaux [8]. The remaining 10% to 15% of cases [15, 26] will experience a slow accumulation of neurological problems from the outset, with occasional plateaux and temporary minor improvements (primary progressive form) [8].

1.2 Epidemiology

The exact causes of multiple sclerosis are not fully known [11, 15, 26]. We do know that the illness is more prevalent in temperate climates and that its incidence increases with distance from the equator [6]. In Canada, the risk of having MS is one of the highest in the world; in 2000-01, it was 0.24% [1]. The Multiple Sclerosis Society of Canada estimates that 55,000 to 75,000 Canadians have the disease [26]. MS is usually diagnosed between the age of 20 and 40, but the illness may appear in people of all ages [16, 26]. The risk is higher for women than it is for men [22, 26].

1.3 General symptoms

At the time when MS is diagnosed, it is impossible to predict its severity, evolution or symptoms [15]. The range of symptoms is wide, probably because deterioration can occur anywhere in the central nervous system [6, 15]. The most common symptoms are weakness, fatigue, impaired motor coordination, bladder problems (incontinence), sensory problems (e.g. impaired sensitivity in one or more limbs, impaired sense of joint position) and vision problems [5, 22]. A person with MS may also show gait impairments; spasticity or low muscle tone; intentional tremor; slowing of fine movements; spasms; pain; dizziness or vertigo; loss of balance; speech problems; facial paralysis; cognitive symptoms; psychological symptoms [5, 6, 15, 22, 26].
2. Ocular and Oculomotor Manifestations

Because multiple sclerosis may affect any region of the central nervous system, it can also cause deficits in the ocular system. Vision impairments are frequent in this population. In the questionnaire-based study conducted by Ford et al. (2001) and that of Lauer and Firnhaber (1987, cited by Richards et al., 2002), half of the people with MS reported vision problems [5, 22]. Another questionnaire survey, published in 1998 by the MS Society of London (cited by Richards et al., 2002), found that 20% of respondents with MS regarded their vision problems as some of the worst symptoms of their illness.

2.1 Optic neuritis

Optic neuritis or the presence of vision symptoms is the first sign of MS in 14% to 27% of cases [6, 10, 14]. Optic neuritis is a demyelinating inflammation of the optic nerve, characterized by reduction or loss of vision in one eye that progresses for about a week [6, 18]. The optic nerve is affected in some 2 out of 3 MS cases (Warner & Lessell, 1994, cited by Gizzi & Kastner, 2000). The resulting visual impairment in the affected eye is typically caused by central scotoma, often in the temporal field [6, 18]. Optic neuritis also causes a total or relative afferent pupil defect1 [6, 10].

The resulting loss of visual acuity (VA) is very variable. A study by the Optic Neuritis Study Group (1991) found that an equal proportion of 448 subjects with a first acute episode of optic neuritis (within the last 8 days) had mild (35%), moderate (29%) or severe (36%) loss of visual acuity [18]. A variety of visual field deficits is also present and optic disc swelling is often observed [10, 18]. Vision loss is usually accompanied by ipsilateral pain in or behind the eye that is worse during eye movements, whether or not the optic disc appears normal or swollen [6, 10, 18].

Because the visual field deficit tends to involve the macula, colour perception and contrast sensitivity impairment are common and may be permanent [6, 10, 18]. Phosphenes (luminous impression occurring when the retina undergoes nonluminous stimulation) may be precipitated by eye movements [10]. In addition, because having only one eye affected results in asymmetry between the eyes in the perception of luminosity and contrast, the optic neuritis may cause an illusion of depth when the person looks at a moving object (Pulfrich effect); they may, for example, be somewhat disoriented in moving traffic [6, 10].

In 90% of cases of optic neuritis, MS-related or not, spontaneous recovery of vision usually occurs with visual acuity of at least 20/40 after the recovery period [19]. This period generally lasts 6 to 12 weeks though improvements may be observed up to 6 months or even a year after onset of the episode [6, 14, 26]. In some cases, however, there is no improvement in vision.

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1 Total afferent pupillary defect: Neither of the pupils contracts in response to a light shone in the affected eye, but both pupils contract if the beam of light is directed at the healthy eye. Relative afferent pupillary defect: During examination, a light is shone in each eye in turn. While the beam moves from the affected eye to the healthy eye, the pupil of the affected eye partially dilates instead of remaining constricted (reduction of afferent signal) [5].
The long-term risks of recurring optic neuritis and permanent damage are twofold higher in people with MS than in others. In a follow-up study by the Optic Neuritis Study Group (2004), 48% of the 148 individuals with MS who had experienced a first episode of optic neuritis 10 years earlier, suffered a recurrence of optic neuritis in one eye during this period (16% in the eye first affected, 19% in the other eye and 13% in both eyes) [19]. During the follow-up examination (10 years after the initial episode), a high proportion had abnormal visual function in the eye first affected, in terms of visual acuity (40%; < 20/20), contrast sensitivity (39%; < 1.65 log units) or visual field (33%; < -3.00 db). Loss of visual function was also measured in the other eye, for 22% to 26% of the subjects, depending on the visual function assessed [19]. This same study showed that a visual impairment may result from episodes of optic neuritis, as shown by the fact that among the total 319 subjects with and without MS, visual acuity in the affected eye was between 20/40 and 20/200 in 5% of cases and lower than 20/200 in 3%.

Symptoms may briefly recur in the affected eye when body temperature rises (e.g. during heat exposure, vigorous exercise, hot bath) [6, 10]. However, this is rare.

Medical treatment of optic neuritis may shorten recovery time but has no impact on the end result, i.e. the extent of possible sequelae [20].

2.2 Defective colour discrimination

Even if they have not had optic neuritis, individuals with MS present progressive impairment in colour discrimination as they grow older. This worsens nearly twice as fast as in people without MS [12]. Sensitivity to colours in the red-green and blue-yellow axes is diminished.

2.3 Visual field damage

Visual field defect may be caused by the presence of demyelinating lesions in the optic chiasm (bitemporal hemianopsia), in the optic tract (homonymous hemianopsia) or in an optic radiation (hemianopsia or quadrantanopsia). Retrochiasmal lesions are relatively rare [6].

Recuperation following the MS episode is variable. It depends on the size of the initial visual field defect; the more extensive the lesion, the more residual defect is likely to be [6].

2.4 Nystagmus

Various types of nystagmus\(^2\) may occur in MS people. The most common are eccentric gaze nystagmus (rebound nystagmus comprising a slow phase and fast phase) and pendular nystagmus (same eye motion velocity in both directions, low amplitude and high frequency) [6, 24]. Although both usually occur in a horizontal plane, they may also be vertical.

Other types of nystagmus may also be present, e.g. dissociated vertical nystagmus (one eye oscillating vertically); gaze-evoked nystagmus (occurring when gaze position changes; present with cerebellar lesions); vestibular nystagmus (may be of fixed direction or may change direction depending on head and trunk position; causes vertigo; is commonly associated with vestibular lesions) [6]. When nystagmus is secondary to a cerebellar or brainstem lesion, its velocity increases during attempts to fixate; this makes reading difficult or impossible [6].

Nystagmus may cause vertigo, blurred vision due to the inability to fixate, and oscillopsia (oscillating vision where objects seem to swing, move back and forth, etc.) [6, 24]. This last symptom may cause unstable gait [14]. Vertigo may disappear after a few days or weeks, but not nystagmus [6]. The Multiple Sclerosis International Federation (2007) reports that unlike other visual symptoms associated with MS, nystagmus is permanent in 70% of cases [13]. However, Rougier & Tilikete (2008) mention the possibility of pharmacological treatment to alleviate or eliminate nystagmus.

2.5 Internuclear ophthalmoplegia

The oculomotor deficit most often associated with MS is internuclear ophthalmoplegia (INO). This is present when there is damage to the posterior longitudinal bundle (medial longitudinal fasciculus) [6]. This bundle (fasciculus) connect various nuclei of oculomotor nerves; it ensures proper functioning of the oculomotor muscles that coordinate abduction of one eye with adduction of the other during lateral gaze in one direction [2, 6]. For example, when you look to the right, your right eye is in abduction (temporal direction) while your left eye is in adduction (nasal direction). A lesion in the bundle fibres results in impaired adduction, with severity ranging from slowing of the movement to paralysis, associated with nystagmus in the abductive eye [2, 6, 24].

INO is rarely symptomatic [24]. The sign most commonly reported is diplopia [6, 14, 24], which may be triggered or exacerbated by fatigue, increased body temperature or stress [13]. There may also be impaired interocular alignment, which may cause fatigue during reading or a phenomenon of visual confusion [6, 24]. A study by Bolanos, Lozano & Cantu (2004), cited by Rougier & Tilikete (2008), found that INO persists for over a year in half the individuals suffering from it. While medication appears to accelerate recovery from diplopia, it does not affect the degree of recovery [14].

2.6 Ocular dysmetria and gaze paresis

Lesions affecting cerebellar white matter are fairly common in persons with MS and may have major consequences for oculomotor performance [6]. Cerebellar pathways are responsible for integrating gaze velocity towards a stationary object and calculating the eye correction movement required to attain the target [6]. Unilateral slowing of smooth ocular pursuit, which translates into jerky pursuit of the target, is commonly observed in MS individuals [6]. However, they are rarely conscious of it, except that they may read more slowly [6]. Related symptoms include blurred vision and oscillopsia during gaze in the affected direction [6].
3. Possible drug side-effects on vision

Certain medications used to treat MS symptoms may have side-effects that affect vision. Gizzi & Kastner (2000) cite a few examples: anti-spasmodics may cause gaze-evoked nystagmus or slowing of visual pursuit; medication for bladder problems may cause blurred vision; treatment with tricyclic antidepressants may cause pupillary accommodation problems; anti-fatigue medication may be associated with nystagmus.

4. Visual impairment interventions

4.1 Global assessment of needs

Quality of life for persons with MS declines as the severity of their symptoms increases [5, 7, 19]. Multiple symptoms can make daily tasks very difficult and vision problems simply add to other disabilities [9]. For example, in a study by Cervera-Deval et al. (1994), vision problems were associated with limitations as regards personal care, transport and social activities [3]. Furthermore, a study by Rudick, Miller et al. (1992), cited by Hakim, Bakheit et al. (2000), found that MS-related vision impairments may lead to job loss.

Vision rehabilitation for individuals with MS needs to take into account the physical and psychosocial impacts of the illness. Among other factors, the approach will depend on whether or not the person has motor disabilities (e.g. weakness, paralysis, spasticity, tremor, dexterity problems) or cognitive problems, and on how rapidly these symptoms evolve. Clinicians also need to consider whether vision loss is permanent or temporary, the impact of visual field loss on near and distant tasks, the capacity to maintain binocularity following the development of strabism or nystagmus, and hand-eye coordination impairment, as applicable [6]. Given the diversity of possible symptoms, vision rehabilitation interventions are performed on a case by case basis. The articles by Gizzi & Kastner (2000) and Perez et al. (2005) describe various case histories [6, 21].

4.2 Vision rehabilitation interventions

As a rule, magnifying, enhancing contrast and enlarging the visual field should all be considered when devising an intervention plan in vision rehabilitation; the person’s physical and cognitive problems should also be taken into account [6].

It is important to evaluate whether the individual is binocular, whether they can fix their gaze and whether they are photophobic (abnormally sensitive to light) [6]. Individuals with persistent diplopia may prefer to cover one eye when performing activities for short periods, though this is not recommended for long periods; wearing corrective lenses with prisms also appears to help [14]. Magnification may also be helpful in certain cases of nystagmus or visual field impairment. Compensatory techniques for nystagmus can be taught (e.g. rotating the head to take advantage of the null point) and a monocular vision aid such as a telescope may diminish gaze-evoked nystagmus [6]. For persons with oscillopsia, prismatic lenses may be indicated, as may magnification and vision aids [14].
Reduced visual acuity or loss of central vision can be managed with magnification and the appropriate vision aid [6]. Some people find a hand magnifier is sufficient; if the person is unable to hold it and move it around smoothly, a stand magnifier may be indicated [6]. In other cases, an electronic magnifier (e.g. closed circuit television) may be necessary. It is then important to assess whether the person is able to grasp the viewing table and move it around smoothly. If not, would an adaptive device to help them hold it be required? Would a motorized table be more appropriate? Can they use their legs to move the table using a special adaptive device? [6]

Also to be considered is computer access. This may significantly enhance the person’s autonomy and social integration, especially if in addition to vision deficits, the person has upper limb motor disability. For some people, the computer is not only their sole means of reading and writing; it also enables them to perform many other activities such as making telephone calls and controlling their environment [4]. However, making computer technology accessible to people with vision and motor impairments can pose a major challenge for rehabilitation clinicians. This is where interdisciplinary work, in this case close collaboration between the visual rehabilitation specialist and the occupational therapist, comes into its own. According to D’Amour, Boulé & Cyr (2008), the evaluation of solutions involves choosing equipment, software and compensatory strategies appropriate to the specific needs and capacities of the individual. A range of choices is available for adaptation to accommodate motor disabilities. With vision problems, using screen magnifier or reading software is often necessary. Compatibility between equipment and software may be a challenge and require a compromise solution [4]. In order to use these adaptations, cognitive and motor learning will clearly have to be accomplished by the person, for whom the strategies are often linked more to the visual impairment [4]. While access to a computer may be challenging for a severely disabled person, the success of interventions will depend, among other things, on his cognitive capacities and motivation; support from his or her environment and the creativity of clinicians are other important factors [4]. For an MS individual facing visual and motor impairments, adapted communication can open the door to new activities, maintained or renewed social integration and better quality of life – in other words, make a real difference [4].

4.3 Orientation and mobility

In the MS population, falls are not unusual; their frequency ranges between 31 and 65% [17]. Vision factors need to be considered in relation to falls. Certain persons with MS report that diplopia and unreliable eye-movement coordination affect their balance [17]. We should also bear in mind that residual vision symptoms of an optical neuritis episode can affect the person’s balance and risk of falling, especially if they are elderly [27].

Moreover, it is important to remember that people with MS may have diminished lower-limb proprioception (reduced perception of the position and motion of limbs in space); compensatory use of vision is thus an important factor in preventing postural oscillation and loss of balance, and ensuring that the risk of falling does not increase [27]. Visual capacity therefore has to be taken into account.
MS may also cause peripheral neuropathy\(^3\), which can affect balance skills and reception of tactile cues. In people who also have visual impairment, neuropathy in the hands may affect their ability to use a long cane; numbness or tingling in the feet may make it difficult for them to sense where they are in space, detect the edge of a step or recognize changes in ground texture or incline [23]. According to Riddering (2008), a support cane or walker may provide the necessary support to prevent falling and enable the person to obtain additional tactile information from the environment. Compensatory strategies and environmental modifications may also be required to assure maximum safety [23].

5. Psychological and cognitive impacts of MS and possible consequences for vision rehabilitation

Rehabilitation of people with multiple sclerosis with a related vision problem can be challenging. Because MS is a multi-symptom illness, these persons are often overwhelmed by their physical disabilities and make them a priority; as a result, they postpone vision interventions unless their visual impairments become major [13]. Deterioration in their physical condition may also affect their emotional state and their receptiveness to vision rehabilitation [6]. Depression may also interfere with the readiness to accept vision aids and use them [6]. The person may be frustrated or disappointed if visual impairments persist or are difficult to resolve due to other incapacities [13]. Their reaction may include aggressiveness, rejection or avoidance for as long as they refuse to acknowledge the extent and severity of their losses (e.g. visual) [25].

The fact that MS is a degenerative disease that is difficult to predict or manage may cause depression and psychological suffering. Lifetime prevalence of a depressive episode in MS appears to vary between 25% and 50%, which is nearly three times higher than in the general population [15, 25, 26]. Depression may occur as a reaction to the diagnosis, or at any stage of the illness [15, 26]. During the 12 months following a period in which symptoms worsen, a person may feel powerless and find it hard to motivate themselves to make an effort, particularly if they feel they will in any case have to repeat the process [25]. Depression may also be a consequence of brain lesions [15, 25]. With time, the individual may develop flat affect (not appearing to feel anything) or labile mood (a tendency to experience sudden, temporary mood changes, with bouts of uncontrollable and inappropriate laughter or weeping) [25, 26]. In severe cases, the “one step forward, two steps back” aspect coupled with the difficulty of predicting the future may be discouraging, not only for the MS person and their family but also for professionals [25].

Fatigue is probably one of the most disabling and frustrating of MS symptoms [15], and varies widely at different times and from one person to another. It should be taken into account even if it is not explicitly reported by the individual [15]. MS may cause loss of

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\(^3\) Peripheral neuropathy: a disease or degenerative state (as polyneuropathy) of the peripheral nerves in which motor, sensory, or vasomotor nerve fibers may be affected and which is marked by muscle weakness and atrophy, pain, and numbness. Retrieved on October 25th 2001, in MedlinePlus Merriam Webster Dictionary. http://www.merriam-webster.com/medlineplus/Peripheral%20neuropathy
function that is initially imperceptible before increasing to noticeable levels. A functional magnetic resonance imaging study by Wallin et al, 2006 (cited by Segal, 2007) demonstrated that some MS individuals, compared to others, use their brain far more extensively for mental tasks; this may contribute to some of the fatigue observed, although the person is unaware of the additional workload imposed on them [25]. The slow degenerative process can affect not only the body but also the mind.

Cognitive problems are present to varying degrees in 40% to 50% of MS cases [15, 26]. Various cognitive spheres may be affected (perception, judgment, initiative, risk awareness, concentration, memory, attention, empathy, ability to make decisions, etc.) [5, 7, 15, 26]. One of the problems for clinicians is the difficulty of gauging whether or not the individual’s attitude is partly attributable to MS. Because neurological damage may be diffuse, certain cognitive functions may remain intact while others are affected. A person may remember an appointment but be unable to remember that they have not been walking or seeing properly for the last few months [25]. In some cases, concentration or memory problems combined with vision deficits can make events difficult to interpret. If the MS person is looking for an object and says “I know it was there a few minutes ago”, maybe the object has not been moved but the person can no longer see it because of their visual deficit and memory problems; they then start thinking someone has moved the object [13]. Such situations easily become confusing.

6. Conclusion

Vision problems are common in people with multiple sclerosis but they generally recover quite well following a flare-up. However, as attacks accumulate, vision and oculomotor problems may persist. Adding to the physical, cognitive and psychological symptoms, they may further complicate the overall picture and contribute to a significant decline in functional capacity and quality of life. Vision rehabilitation may then be required. When we assess an individual with MS and establish the intervention plan, the wide diversity of possible symptoms make it critically important to use a holistic, interdisciplinary approach in order to address the person’s needs. This will ensure that interventions achieve optimal impact.
7. References


